NEUROMUSCULAR DISEASE

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This supplement tackles the lower levels of the nervous system, the peripheral nerves, the neuromuscular junction, and muscle diseases. Motor neurone disease/amyotrophic lateral sclerosis, a disorder of both upper and lower motor neurones, will be considered in a later supplement.

There are many diseases of nerve and muscle; a few are common, many are rare, a situation that often leaves neurologists with uncertainty as to how extensively to investigate their patients. Some of these conditions are treatable, though in none is this simple. Here we have chosen to emphasise the practical approach to the assessment and management of patients who present with neuromuscular symptoms.

Hugh Willison and John Winer outline their clinical approach to evaluating and investigating peripheral neuropathies, the evaluation of genetically determined neuromuscular disease having been discussed by Mike Hanna and Mary Reilly in the recent issue of *Neurology in Practice* on neurogenetics (see below). When a diagnosis of an inflammatory neuropathy is made there are a number of therapeutic options; Robert Hadden and Richard Hughes review what is available and the evidence for how and when it is used. Many patients with neuropathies are found to have diabetes. This is usually the first step in their evaluation causation and Gareth Llewellyn explores the types of diabetic neuropathies and their management. Focal peripheral neuropathies are very common, on the orthopaedic border of neurology, and Geraint Fuller outlines a practical approach.

While muscular symptoms are commonly complained of, only infrequently do they reflect significant muscular disease; Richard Petty discusses the symptomatic approach to evaluating patients with possible muscle diseases. David Hilton-Jones provides insights into the rare but difficult management of inflammatory muscle diseases. Bridging "the gap", Marguerite Hill discusses disorders of the neuromuscular junction, their diagnosis and management.

Much of the management of neuromuscular disease has evolved without being subjected to evaluation in clinical trials. Kate Jewitt and Richard Hughes provide a Cochrane view of how this is changing.

The route map below, as always, tries to highlight some useful references.

KEY REFERENCES

Genetic neuromuscular disease

- Genetic neuromuscular disease. Reilly MM, Hanna MG. *J Neurol Neurosurg Psychiatry* 2002;**73**(suppl II):ii12–21.

Clinical and laboratory approach to genetic neuromuscular disease.

Neuropathies

Guillain-Barré syndrome (GBS)

- Guillain-Barré syndrome. Hahn AF. *Lancet* 1998;**352**:635–41.
- Pathogenesis of Guillain-Barré syndrome. Hughes RAC, Hadden RDM, Gregson NA, Smith KJ. *J Neuroimmunol* 1999;**100**:74–97.

A detailed review on the cellular and antibody mediated mechanisms of pathogenesis of Guillain-Barré syndrome.

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)

- Chronic inflammatory demyelinating polyradiculoneuropathy. Pollard JD. *Curr Opin Neurol* 2002;**15**:279–83.

Multifocal motor neuropathy (MMN)

- Multifocal motor neuropathy. Nobile-Orazio E. *J Neuroimmunol* 2001;**115**:4–18.

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Paraproteinaemic neuropathy

- Neuropathies associated with paraproteinemia. Ropper AH, Gorson KC. *N Engl J Med* 1998;**338**:1601–7.
- Neuropathy and monoclonal gammopathy. Nobile-Orazio E, Carpo M. *Curr Opin Neurol* 2001;**14**:615–20.

Intravenous immunoglobulin

- Association of British Neurologists guidelines for the use of intravenous immunoglobulin in neurological diseases (March 2002).

Useful information on indications, adverse effects, monitoring, consent and differences between brands. Text may be downloaded from www.theabn.org

- Mechanism of action of intravenous immunoglobulin and therapeutic considerations in the treatment of autoimmune neurologic diseases. Dalakas MC. *Neurology* 1998;**51**(6 suppl 5):S2–8.

A review of probable mechanisms of action of intravenous immunoglobulin.

Focal peripheral neuropathies

- *Mononeuropathies: examination, diagnosis and treatment*. Staal A, van Gijn J, Spaans F. London: WB Saunders, 1999.
- *Focal peripheral neuropathies*, 3rd ed. Stewart JD. Lippincott, Williams, Wilkins, 2000.

Two excellent summaries of all focal neuropathies. Know where to find a copy of one of these references.

Muscle disease

- *Disorders of voluntary muscle*, 7th ed. Karpati G, Hilton-Jones D, Griggs RC, eds. Cambridge University Press, 2001.

A comprehensive source of information about muscle diseases.

Erratum

Schott JM, Fox NC, Rossor MN. Genetics of the dementias. *J Neurol Neurosurg Psychiatry* 2002;**73**(suppl II):ii27–31. In this article, it was stated that the error size of the prion protein gene was 100 bp when it is approximately 750 bp.